Case reports

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Bacterial endocarditis of mitral valve in Marfan syndrome

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An unusual case of bacterial endocarditis in Marfan syndrome is reported. A review of the published reports revealed 21 previously reported cases. The aortic valve, though commonly abnormal in Marfan syndrome, was rarely involved by the endocarditis. In contrast, the mitral valve was the favoured site of infection in these patients. Our own patient had staphylococcal endocarditis of both the aortic and mitral valves, the only such combination of infecting organism and sites in the published reports to date. The low incidence of aortic valve involvement remains unexplained. There is extreme mortality in Marfan patients affected by endocarditis, and, it seems, only one documented cure in the entire published reports.

The Marfan syndrome, a heritable disorder of connective tissue (Marfan, 1896), is characterized by variable abnormalities of skeletal, ocular, and cardiovascular systems. Cardiovascular involvement is important because of its prognostic significance (Murdoch et al., 1972). The most commonly recognized cardiovascular defects are aortic dilatation and its complications, aortic regurgitation and dissecting aneurysm. Deformity of the mitral valve in a patient with Marfan syndrome was first described by Salle (1912). Subsequently, others (Traisman and Johnson, 1954; McKusick, 1966) emphasized that mitral valve abnormalities may occur in patients with this syndrome. Few cases of bacterial endocarditis in Marfan syndrome have been reported, despite the known susceptibility of diseased valves to such infection (Hiejima et al., 1968). Furthermore, it is curious that when bacterial endocarditis occurs it almost always involves the relatively uncommon mitral abnormalities rather than the more common lesions of the aortic valve (Di Matteo et al., 1971; McKusick, 1972).

In this report we describe our experience with a patient who had typical Marfan syndrome and died of acute staphylococcal endocarditis involving both the aortic and mitral valves.

Case report

A 14-year-old white boy was admitted to the Jersey City Medical Center on 8 August 1971 in a semiconscious state. The day before admission he had fever, severe headache, two episodes of projectile vomiting, and gradually became drowsy. There was no history of convulsions, head injury, ear discharge, recent dental treatment, or narcotics use. A month before admission, the patient had an upper respiratory infection, for which he received symptomatic treatment.

Six years previously he was treated in another hospital for pneumonia, and was told that he had a heart murmur, but no diagnostic procedures were performed. There were no other major illnesses or operations.

Family history

The patient's mother was alive and well. His father had Marfan syndrome, and died suddenly at home in 1971, after having severe chest pain for several hours. No necropsy was performed. The patient had 3 brothers and 2 sisters. All but one (a sister) were said to have Marfan syndrome.

Physical examination

On admission the patient was stuporous, restless, and dehydrated. His temperature was 40.5°C, pulse 148 a minute and regular respirations 40 a minute, and blood pressure 130/60 mmHg. His height was 184 cm and his span was 207 cm. Extreme arachnodactyly was present. There were no skull deformities. He had a high arched palate. A pectus carinatum deformity was present on an elongated chest. The pupils were normal, reacted well to light, and showed no quivering of the irises. Slit-lamp examination was not done. The fundi were normal. There was slight neck stiffness. The deep tendon reflexes were depressed throughout, and the Babinski sign was elicited on the left. There were no other abnormal reflexes, sensory deficits, or motor weaknesses. Examination of the heart showed sinus tachycardia. The apex
beat was in the fifth intercostal space in the midclavicular line, with sounds of normal quality. There was a pansystolic grade 3/6 murmur, best heard at the apex, conducted towards the left axilla. No other murmurs were heard. All peripheral pulses were easily felt. The lungs were clear and the abdomen normal. There was no hepatosplenomegaly.

**Laboratory investigations**
White cell count 13,000/mm³, 74 per cent segmented neutrophils, 15 per cent stab forms, 10 per cent lymphocytes, 1 per cent monocytes. Haematocrit, haemoglobin, serum electrolytes, and blood urea nitrogen normal. Lumbar puncture: initial pressure 250 mm (H₂O), final pressure 170 mm; protein 10 mg/100 ml, sugar 110 mg/100 ml, cell count 60/mm³ with 90 per cent polymorphonuclears. Urinalysis: specific gravity 1.021, 4+ albumin, many red blood cells and epithelial casts, few white cells. Five blood cultures, reported after death: pure cultures of coagulase positive *Staphylococcus aureus*. Chest x-ray showed moderate dilatation of ascending aorta, clear lung fields, normal cardiac contour. Electrocardiogram was suggestive of left ventricular and right atrial enlargement.

**Hospital course**
A provisional diagnosis of Marfan syndrome with meningitis was made, and treatment was started with intravenous ampicillin (multiple blood cultures were drawn). However, four hours after admission, he lapsed into coma, and died 32 hours later.

**Necropsy**
The pertinent postmortem findings were in the heart, brain, liver, kidneys, and pituitary, and adrenal glands. The heart was enlarged and weighed 370 g. The left

<table>
<thead>
<tr>
<th>Reference</th>
<th>Evidence of bacterial endocarditis</th>
<th>Organism</th>
<th>Valve affected</th>
<th>Operation and/or necropsy</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Olcott (1940)</td>
<td>Heart blood culture</td>
<td><em>Esch. coli</em></td>
<td>Mitral</td>
<td>Yes</td>
<td>A case of arachnodactyly and aneurysmal dilatation of thoracic aorta</td>
</tr>
<tr>
<td>2) Vivas-Salas and Sanson (1948)</td>
<td>Histological examination of mitral valve</td>
<td>Gram-positive cocci</td>
<td>Mitral</td>
<td>Yes</td>
<td>Positive family history</td>
</tr>
<tr>
<td>3) Schorr, Braun, and Wildman (1951)</td>
<td>Recurrent fever, responding to antibiotics; blood culture negative</td>
<td>Not isolated</td>
<td>Unknown</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>4) McKusick (1955)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Mitral</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>5) Tolbert and Birchall (1956)</td>
<td>Blood culture</td>
<td>Haemolytic streptococci</td>
<td>Mitral, ventricular septal defect, persistent ductus arteriosus</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>6) Pappas, Mason, and Denton (1957) (Case 2)</td>
<td>History of subacute bacterial endocarditis</td>
<td>Streptococci</td>
<td>Aortic</td>
<td>Yes</td>
<td>No bacterial vegetations; perforation in aortic cusp from bacterial endocarditis</td>
</tr>
<tr>
<td>7) Hirosawa et al. (1957)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Ventricular septal defect</td>
<td>No</td>
<td>History of acute rheumatic fever at age 10 years</td>
</tr>
<tr>
<td>8) Miller and Pearson (1959)</td>
<td>Mitral valve showed vegetations</td>
<td>Unknown</td>
<td>Mitral</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>9) Sinclair, Kitchen, and Turner (1960) (Case 15)</td>
<td>Past history of subacute bacterial endocarditis treated successfully</td>
<td>Unknown</td>
<td>Unknown (basal systolic murmur)</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>10) Bowers and Lim (1962)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Eisenmenger's complex</td>
<td>No</td>
<td>Phenotypically Marfan syndrome with intracranial calcification and history of maternal rubella; probably first documented cure of subacute bacterial endocarditis</td>
</tr>
<tr>
<td>11) Wunsch et al. (1965)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Mitral</td>
<td>Yes</td>
<td>Patient's father probably had Marfan syndrome</td>
</tr>
</tbody>
</table>
TABLE (Cont’d)

<table>
<thead>
<tr>
<th>Reference</th>
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<th>Organism</th>
<th>Value affected</th>
<th>Operation and/or necropsy</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>12) Maekawa (1965)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Ventricular septal defect</td>
<td>Yes</td>
<td>Normal physical appearance, floppy-valve syndrome</td>
</tr>
<tr>
<td>13) Read, Thal, and Wendt (1965)</td>
<td>History of pneumococcal endocarditis at age 7 years</td>
<td>Unknown</td>
<td>Mitral</td>
<td>Yes</td>
<td>Moderate hypermobility of joints and scoliosis, floppy-valve syndrome</td>
</tr>
<tr>
<td>14) Read et al. (1965) (T.J.)</td>
<td>History of streptococcal endocarditis at age 23 years</td>
<td>Unknown</td>
<td>Mitral</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>15) Shiina et al. (1966)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Mitral</td>
<td>Yes</td>
<td>Positive family history; mild pulmonary stenosis; abnormality of aortic valve on angiocardiography</td>
</tr>
<tr>
<td>16) Keech et al. (1966) (T.J.Jr.)</td>
<td>Clinically probable (repeated blood cultures negative)</td>
<td>Unknown</td>
<td>Unknown</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>17) Iwase (1967)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Ventricular septal defect</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>18) Cohen and Kaye (1967)</td>
<td>Blood culture</td>
<td><em>Staph. aureus</em></td>
<td>Mitral</td>
<td>Yes</td>
<td>Heroin addict; no predisposing lesion of mitral valve; but histology consistent with bacterial endocarditis</td>
</tr>
<tr>
<td>19) Hiejima et al. (1968) (Case 1)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Mitral</td>
<td>Yes</td>
<td>Myopia in sibs and mother</td>
</tr>
<tr>
<td>20) Aslam et al. (1970)</td>
<td>Blood culture</td>
<td>Aspergillus</td>
<td>Aortic</td>
<td>Yes</td>
<td>Family history suggestive of Marfan; postoperative endocarditis; aortic valve showed thin fenestration</td>
</tr>
<tr>
<td>21) Di Matteo et al. (1971)</td>
<td>Blood culture</td>
<td><em>Strep. viridans</em></td>
<td>Mitral</td>
<td>Yes</td>
<td>Clinically forme fruste Marfan</td>
</tr>
</tbody>
</table>

ventricular wall measured 1–2 cm and the right ventricular wall 0–3 cm. The aortic and mitral valvular circumferences were enlarged: 7.5 cm and 11 cm, respectively. Both valves were thickened at their free borders and showed occasional pinpoint greyish elevations. No gross lesions could be recognized in the myocardium. The ascending aorta was moderately dilated. Microscopical examination of the mitral and aortic valves showed vegetations populated by Gram-positive cocci. At the bases, remote from any exudate, there was prominent mucinous degeneration of the ground substance. This was more noticeable in the aortic valve, where there was a minute linear defect partially filled by a few red blood cells. There were numerous micro-abscesses within the myocardium, liver, kidney, brain, pituitary, and adrenal glands. (Postmortem blood culture from the heart was positive for *Staphylococcus aureus*, coagulase positive.)

Discussion

The first case of bacterial endocarditis in Marfan syndrome was reported by Olcott (1940). While isolated cases continue to be reported, the exact incidence of endocarditis among patients with Marfan syndrome cannot be determined (Wunsch, Steinmetz, and Fisch, 1965; Iwase, 1967; Di Matteo et al., 1971). Our own survey revealed 21 adequately documented cases, the important features of which are summarized in the Table.

Of the 21 patients, the infecting organism was identified in 15. It was not demonstrated in 6 cases, but other supporting evidence for the diagnosis of bacterial endocarditis was available. Streptococci were isolated in 11, of which 9 were *Streptococcus viridans*. A variety of organisms was isolated in 4 patients. *Staphylococcus aureus*, which was the offending organism in our patient, was present in only one other recorded case of endocarditis in Marfan syndrome (Cohen and Kaye, 1967).

Clinical examination pointed to the site of the
underlying cardiac lesion in 18 patients. This location was confirmed by surgery or necropsy in 15. Congenital malformations were present in 5. The mitral valve was involved alone 11 times and the aortic valve only twice. Thus, the mitral valve was the site of infection in almost two-thirds of these patients. Considering the well-known susceptibility of aortic valve lesions to bacterial infection, and the predominant involvement of the aortic valve in Marfan syndrome, this finding was unexpected. Why the aortic valve is so much less vulnerable than the mitral valve is unexplained. To our knowledge, our patient is the first showing histological evidence of bacterial endocarditis affecting both the aortic and mitral valves.

The Marfan syndrome with bacterial endocarditis has a dismal prognosis, one that is probably worse than that of patients with endocarditis complicating other forms of heart disease. We were able to find only one documented cure in the published reports. A high degree of suspicion, early diagnosis, and the prompt institution of treatment may improve the otherwise grave prognosis of these patients.

References
Vivas-Salas, E., and Sanson, R. E. (1948). Sindrome de Marfan, sin cardiopatia congenita y con endocarditis lenta confirmada por la autopsia. Archivos del Instituto de Cardiología de Mexico, 18, 217.

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